When Is It Time To Do Surgery from a Pulmonary Perspective

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Disclosures

- Pediatric Pulmonary Section Editor of UpToDate
What do we (think we) Know?

- The 3-dimensional spine/thoracic deformity leads to lung impairment that worsens as the deformity worsens.
- Pre-operative lung function is a major determinant of post-operative lung function (long and short term).
- Lung function after insertion of growth friendly distraction devices does not improve in most children but allows further lung function growth (in liters).
- Co-morbid states, such as neuromuscular weakness often determine the pulmonary outcome of children despite the scoliosis repair.
Seattle-Philadelphia-San Antonio: First FVC (% predicted) *

CSSG Registry: 54% of 3,968 patients > 5 years old at presentation
# Classification for Early Onset Scoliosis + Pulmonary Data

<table>
<thead>
<tr>
<th>Age</th>
<th>Etiology</th>
<th>Major Curve Angle</th>
<th>Kyphosis</th>
<th>APR Modifier</th>
<th>FVC</th>
<th>PFT Modifier</th>
</tr>
</thead>
<tbody>
<tr>
<td>Continuous Prefix</td>
<td><strong>Congenital/Structural</strong></td>
<td>1: &lt;20°</td>
<td>(−): &lt;20°</td>
<td>P₀: &lt;10°/yr</td>
<td>&gt;70%</td>
<td>&lt;5%/yr</td>
</tr>
<tr>
<td></td>
<td><strong>neuroMuscular</strong></td>
<td>2: 20-50°</td>
<td>N: 20-50°</td>
<td>P₁: 10-20°/yr</td>
<td>50-70%</td>
<td>5-10%/yr</td>
</tr>
<tr>
<td></td>
<td><strong>Syndromic</strong></td>
<td>3: 51-90°</td>
<td>(+) : &gt;50°</td>
<td>P₂: &gt;20°/yr</td>
<td>30-50%</td>
<td>&gt;10%/yr</td>
</tr>
<tr>
<td></td>
<td><strong>Idiopathic</strong></td>
<td>4: &gt;90°</td>
<td></td>
<td></td>
<td>&lt;3%</td>
<td></td>
</tr>
</tbody>
</table>

**Kyphosis**
- **P₀**: <10°/yr
- **P₁**: 10-20°/yr
- **P₂**: >20°/yr

**PFT Modifier**
- <5%/yr
- 5-10%/yr
- >10%/yr

**FVC**
- >70%
- 50-70%
- 30-50%
- <3%
Spine/Pulmonary Joint Decision Points

- Is there a threshold lung function value where I have to intervene immediately?
- What degree of lung function loss can I tolerate as a spine deformity progresses?
- How often do I follow lung function to assess trends?
- How do I juxtapose this decision with age of surgical preferences, e.g. fusion?
Serial FVC Over a 6 Year Interval

28% Reduction from Pre to LAST

16% Reduction Over 6 Years


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Ages and Treatment Options for Children with EOS

- **<4 years**
  - Non-Surgical
  - Growth Friendly Devices

- **4-6**
  - PFTs

- **8-12**
  - Growth Friendly Devices

- **>12 years**
  - Fusion
Evidence for Earlier Intervention

- VEPTR insertion and expansion (6 years F/U)
  - N=16 <6 y/o; N=7 >6 y/o
  - FVC change per year: <6 y/o: 14.7+/-8%
    >6 y/o: 6.5+/-5.5%

- Vertebral Column Resection (2 year F/U)
  - N=27 children, ages 8-18 years
    - Correlation between age of surgery and change in FVC (not % predicted) r=-0.44; p=.02

Decline in FVC after surgery spine fusion for DMD based on age

\[ N = 56 \]
Speculation

- Earlier *intervention* to prevent deformity progression is better for lung function preservation than later. Intervention does not mean surgery or type of surgery.

- *Ideal timing* of surgical interventions may not coincide with ideal timing to stop lung function decline. How do we decide next steps?

- New methods are needed to *salvage* lung function which is severely impaired at presentation.